

HUNTINGTON NUMBER

103

Neurology. Huntington's Chorea

NEUROGRAPHS

A SERIES OF NEUROLOGICAL
STUDIES, CASES, AND NOTES

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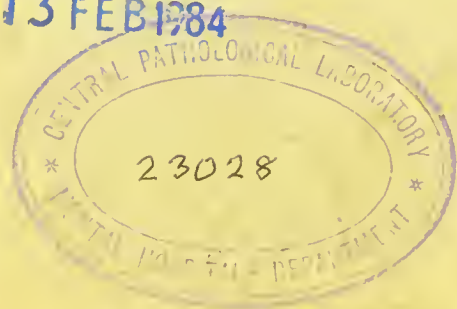
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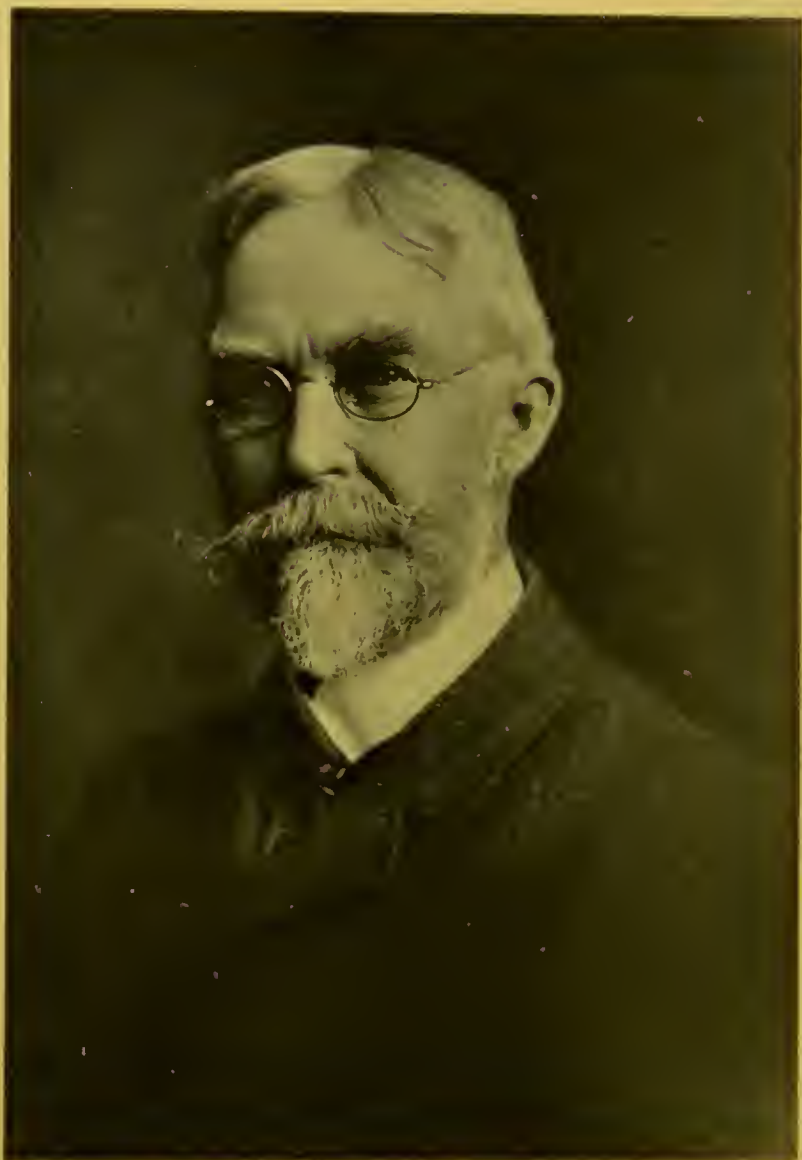


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Frontispiece.



Geo. Huntington

Neurographs

EDITORIAL.

The Huntington Number.—In view of the world-wide interest that has been shown in the subject of hereditary degenerative chorea, of the fact that Dr. Huntington is a native of Long Island and that his cases were described from here, and that comparatively little personal appreciation has been shown him, although fortunately he is still active in the profession, it has seemed worth while to devote this number of NEUROGRAPHS partly to honoring him and partly to what is believed to be a useful clearing-up of the record.

The three contributions from abroad not only carry their own measure of enlightenment, but, coming from the highest European authorities, are a practical expression of the general importance of this subject and constitute a most graceful international tribute.

Huntington's Work.—Our knowledge of this form of chorea dates entirely from the article by Dr. George Huntington, in 1872. There were good reasons why his paper succeeded in drawing general attention to this disorder and in securing for it permanent recognition.

I.—As stated by Eichhorst, "Hereditary chorea of adults was first fully described by Huntington, and is therefore known as Huntington's Chorea."

II.—Huntington was the first to give definitely the location of his cases, and thus positively establish a verifiable record. It is a dictum of experimental medicine as well, that only what is verifiable can be accepted.

III.—The abstracting of his original article by Kussmaul and Nothnagel, in Virchow-Hirsch's *Jahrbuch* for

1872. Thanks to the work of Friedreich on hereditary ataxia, as well as the growing interest in heredity, the time was ripe for its appreciation.

Only work of unusual incisive and wide-reaching interest could attract such a share of attention. What he described had interest both as a disease and in its bearings on questions of heredity. As one corollary it removed the age-limit, demonstrated at a sweep that hereditary factors can become dominant at any period of life.

Such work could not well originate in a laboratory or technical institution. It belongs to what has been considered the newer and exclusively medical field, the scope of progress that remains to the medical man as such.

The Term, "Huntington's Chorea."—Operations, methods, and instruments in considerable number are known by the names of their American originators or describers. But, in the list of diseases very few are thus designated. Neurasthenia may occasionally be termed Beard's disease, or oftener dermatitis herpetiformis may be called Duhring's disease, and a few others may likewise bear a corresponding distinction. Yet no single disease of equal importance is so commonly designated and understood by a native's name. There are arguments against this custom, such as have been strongly urged by my friend, Prof. Wilder, but that is not the point here. It is to the accomplished fact that we refer. However, these things are not altogether accidental, though even then of interest. Many attempts have been made, notably in this country, to elide this word and substitute some more technical designation.* But this country does not have the decid-

*The following list includes some of the more frequently used synonyms: Huntington's Chorea; adult hereditary chorea; chronic progressive chorea; chronic chorea; choreic dementia; degenerative chorea; chorea of the aged; family chorea. Of course these forms are Latinized and rearranged at the will of individual writers.

ing of these matters. And, somehow, despite attacks and without effort on anyone's part to retain the eponym, the form "Huntington's chorea" is continued. In reality it is the most scientific, because the simplest and most distinctive of any name yet applied to this condition and the one that is known and used in medical circles everywhere. When this term is employed, one is aware just what is intended, while no other form has to anyone's mind the same exact and unquestioned delimitation.

There is another favorable side to the use of this. It balances up with the nomen often used for ordinary chorea; so that in Sydenham's chorea and Huntington's chorea we have convenient, yet equal and contrasting designations for the two general types.

For these ample reasons the designation is bound to be preserved. It can be Latinized, as is done abroad, into *chorea Huntingtonii*, and in that form should satisfy the fastidious scientist who is not a pedant.

The Earlier American Observations.—It has been possible by due investigation to work out the heretofore unknown factor in the earlier observations (*v. infra*) of this disorder, *i. e.*, the localities where these were made and the relation of the cases to others. This is a matter, of both historical and scientific interest, a necessary preliminary to the tracing of isolated or collective cases and, in fact, to any comprehensive study of the subject. To accomplish this it was necessary to work out the personal histories of each of the three earlier observers of this disease. Of none of them was anything previously known in this connection beyond his single brief paper. Who they were, what became of them, whence their cases, their scientific responsibility—on all these points no light had been shed even by those who had resurrected their writings. They remained more obscure than the

pre-Columbian discoverers of America, and in fact their relation to the main point much resembles that of the Norsemen to the discovery of America. It now transpires that each had subsequently shown himself to be a man of ability, character, and power of observation. All these facts can best be made clear (and, it is hoped, satisfactorily settled) by giving under separate headings a brief outline of the life of each.

It is a notable fact that all four of the earlier describers were young men, and at the time but just out of medical schools.

The Historic Method in Investigation.—Some method, such as may be suggested by this heading, needs be found for turning to medical and scientific account the huge stores of material that of late years, and in this country at least, have been gathered and more or less systematized by our genealogical enthusiasts. Never and nowhere else in the world has an equal basis of collected material existed. Of course this should be collated with other data. The allied class of facts in *Who's Who* has been made use of in certain ways. Part of the scheme of the present number is aimed in this direction.

In the thirty-six years since Huntington's writing, little, if any knowledge has accumulated regarding the prevention or relief of the trouble he described. Time then to approach the matter on radically different lines. From its most marked feature, heredity, an historical order of investigation comes naturally first to mind. How much it may yield can not be predicted until it has been well tried.

* * * *

To all those who have so courteously contributed articles or so kindly aided with information bearing on the subject, the Editor begs to express appreciation and obligation.

A BIOGRAPHICAL SKETCH OF GEORGE HUNTINGTON, M.D.

By James Macfarlane Winfield, M.D.,

Professor of Diseases of the Skin, Long Island College Hospital,
Brooklyn, New York.

All human achievement in art, science or trade is the result of slow growth and development; "genius is only an infinite capacity for hard work." Some one must have patiently and faithfully toiled to enable others to reap the benefit, and it is an ever-inspiring truth that an effort that ends in apparent disheartening failure in one generation, in the next may be crowned with glorious, and what seems sudden, success.

Personal inspiration is only the fulfillment of the hopes, longings and aspirations of those who have gone before.

If we grant this, then the study of the life of any man who has deciphered for us a new page in Nature's book becomes a fascinating search for the reasons and causes which made it possible for him to read where others had all unwittingly thumbed the leaves.

As we follow a brook up to the spring that gives it life, so we may trace a man's capabilities and characteristics through the blood of those from whom he sprang and find the source of his success.

George Huntington's life is especially interesting from this viewpoint, for his clearly established ancestry, traced back to well-known and gentle English stock, and the carefully kept records of the successes and labors of his immediate ancestors, make it genetically proper that the medical world call chronic hereditary chorea by his name.

In 1633, Simon Huntington, of Norwich, England,

and his wife, Margaret Baret, with their children, sailed for America. The father died on ship-board and the widow and children probably landed at Saybrook, Conn. In 1652, Christopher, the third son, married Ruth Williams, of Windsor, Conn., from which place he moved to Saybrook, and in 1660, with his brother Simon, aided in laying the foundations of the present city of Norwich, Conn.

Christopher, the son of Christopher and Ruth, was born November 1, 1660, and was the first male child born in Norwich. He married Sarah Adgate, of Norwich; their seventh son, John, married Sibil Tracy.

Ezra, the fourth son of John and Sibil, married his cousin Elizabeth Huntington.

Abel, the sixth son of Ezra and Elizabeth, and the grandfather of the subject of our sketch, was born in Norwich, Conn., February 21, 1777.

He studied medicine with Dr. Philemon Tracy, of Norwich, a noted physician in the state. Dr. Abel Huntington married Frances Lee, the daughter of a revolutionary officer. In 1797, they settled in Easthampton, Long Island, and he immediately became the leading physician of that section. That he was an advanced and successful physician and surgeon is attested by the fact that he was the first on Long Island to perform the operation of lithotomy. He was in charge of the retreat for those who had been inoculated for smallpox, and personally prepared and preserved the variola virus. The present Dr. Huntington showed the writer some of these ancient quills hermetically sealed in phials.

Dr. Abel Huntington was one of the first in America to take advantage of Jenner's great discovery. He was active in civil as well as medical affairs, was Presidential Elector in 1820; New York State Senator in 1821;

served two terms as Congressman during the administration of Andrew Jackson, and enjoyed the friendship of that stern, "Old Hickory" President. In 1845 he was appointed Collector of Customs for Sag Harbor, and in 1846 was a member of the committee to revise the constitution of the State of New York.

George Lee Huntington, the only son of Abel and Frances, and the father of the present Dr. George Huntington, was born in Easthampton, L. I., July 15, 1811. He studied medicine with his father; he was a special student of the great Valentine Mott, and later received his degree from the New York University. He joined his father in the practice of medicine in Easthampton where he remained for the rest of his life.

In 1833 he married Mary Hoogland, of New York City; they had two sons who followed the medical profession, the subject of our sketch, and his eldest brother, Dr. Abel Huntington, who was for many years the Medical Director of the New York Life Insurance Company.

When George Huntington was born at Easthampton, Long Island, April 9, 1850, the two great factors in a man's life, heredity and environment, were both in his favor. We have traced the fact that those who went before him were men of intellectual and moral power, educated, broadminded and public spirited; from them the boy inherited the ability to observe accurately and report scientifically what was interesting or unusual in his surroundings.

His environment was quaint and beautiful, and would incline a sensitive, imaginative child to a poetic rather than a practical comprehension of life. He was the son of the much respected doctor of Old Easthampton, a drowsy, secluded village at the extreme

end of Long Island; settled by New Englanders, it has the attractive characteristics of their villages—the long common green, the beautiful well-kept trees, and the comfortable, substantial houses, each with an individual air combined of self-respecting, secluded dignity and kindly hospitality, the latter, however, to be dispensed only to the virtuous and the deserving from a strictly high-bred standpoint; and over all an air of peaceful home life.

Ocean and forest were then the teachers and playground of Easthampton childhood; a charming, soft, dreamy atmosphere hushed noisy activity and restless ambition, and woke the imagination and the faith in legends and the invisible.

Here George Huntington worked and played, dreamed and studied, attending the old "Clinton Academy" of Easthampton.

He obtained his classical education from Mr. John Wallace, a graduate of Edinburgh University, Scotland, and a one time Lord High Sheriff of Edinburgh.

Every good man's life owes much to some woman; *cherchez la femme* is divinely as well as profanely true. Huntington's mother was of a fine old Knickerbocker family, a woman of education and great force of character; and his father's sister, who made a member of the household during his boyhood, was a writer of literary ability, and published several books of poems of merit and charm.

He studied preliminary medicine with his father and was graduated from the College of Physicians and Surgeons, Columbia University, N. Y., in the spring of 1871. His graduation thesis was upon "Opium."

After obtaining his medical degree he returned to Easthampton and assisted his father in practice.



GEORGE HUNTINGTON, M.D.
(IN 1872.)

The families he attended were of old New England stock, and from the natural seclusion in which they lived had often intermarried.

Among them existed a peculiar nervous disease, the medical recognition and scientific description of which has made Dr. Huntington famous.

This disease had been recognized by Dr. Huntington's grandfather, Dr. Abel Huntington. The cases Dr. Huntington describes had been classified by his father, Dr. George Lee Huntington, and he had the benefit of the elder doctors' experience and talent; the manuscript of his original article on chorea was carefully revised by his father, and bears the correctional marks of his pencil.

This article is classic in its accurate, unmistakable characterization of symptoms, and from this word picture any tyro could recognize Huntington's chorea. It is natural that this hopeless disease with its hideous and grotesque symptoms should strike sharply on the artistic, beauty-loving intellect of the young physician and crystallize in a description of photographic, almost living, accuracy.

In 1871, Dr. Huntington went to Pomeroy, Ohio, with the object of settling in practice there; a cousin of his had married a prominent clergyman of Pomeroy who encouraged him to this step.

There he met Mary Elizabeth Hackard, daughter of Judge Martin Hackard, whom he married in 1874.

Dr. Huntington found Pomeroy abundantly supplied with physicians, and as the chance for successful competition seemed small, returned to Easthampton.

In 1874 he settled in La Grangeville, Dutchess County, New York; here he remained until 1901 when ill-health compelled him to go to Asheville, North Carolina.

Here he practiced medicine and joined the state society. In 1903 his health was sufficiently restored to permit him to return to Dutchess County, and he settled at Hopewell Junction, where he is now engaged in general practice.

Dr. Huntington has five children: Charles Gardnier, Edwin Horton, Catherine, Elizabeth and Eleanore; they show the intellectual and musical ability of their heredity.

Dr. Huntington is a man of charming personality, an artist and musician of ability, especially gifted in making excellent pen and ink sketches, and very clever at caricature in which his keen observation and delicate humor find facile expression.

He is a lover and student of nature and her woodcraft secrets, and an ardent devotee of the rod and gun; this, coupled with a retiring, highly refined and idealistic temperament, has led him to prefer the individual and peaceful life of a suburban practitioner to the stress and emulation of a city practice, although, had he chosen the latter, his fine training and education, and his concise and scientific manner of observing and recording would, doubtless, have obtained him brilliant success.

He is an avid reader and student, and keeps well abreast of the rapid advance of medical science.

Dr. Huntington is the health officer of the township of East Fishkill; visiting physician to the Matteawan General Hospital; member of the American Medical Association, 1905—; Tristate Medical Association (Virginia, North and South Carolina), 1903—; Medical Society of the State of New York, 1880—; Medical Association of the State of New York, 1905; member of Buncombe Medical Society, N. C., 1901 to 1905; honorary member of the Brooklyn Society for Neurology, 1898—; member of Dutchess County Medical Society,

1874—; its vice-president in 1887 and president in 1888; he has also been a member of the New York Audobon Society for many years.

HUNTINGTON'S ORIGINAL DESCRIPTION OF THIS FORM OF CHOREA (FROM THE 1872 PRINT).

After giving a general paper on the subject of chorea in childhood, Huntington devoted the latter part of the article entirely to what he termed hereditary chorea. Osler speaks of this description as, "the really graphic account he has given in a few paragraphs of the salient features of the disease." The original text is as follows:

"And now I wish to draw your attention more particularly to a form of the disease which exists, so far as I know, almost exclusively on the east end of Long Island. It is peculiar in itself and seems to obey certain fixed laws. In the first place, let me remark that chorea, as it is commonly known to the profession, and a description of which I have already given, is of exceedingly rare occurrence there. I do not remember a single instance occurring in my father's practice, and I have often heard him say that it was a rare disease and seldom met with by him.

"The *hereditary* chorea, as I shall call it, is confined to certain and fortunately a *few* families, and has been transmitted to them, an heirloom from generations away back in the dim past. It is spoken of by those in whose veins the seeds of the disease are known to exist, with a kind of horror, and not at all alluded to except through dire necessity, when it is mentioned as '*that disorder*.'

"It is attended generally by all the symptoms of common
"chorea, only in an aggravated degree, hardly ever mani-
"festing itself until *adult* or *middle* life, and then coming
"on gradually but surely, increasing by degrees, and
"occupying years in its development, until the hapless
"sufferer is but a quivering wreck of his former self.

"It is as common and is indeed, I believe, *more* common
"among *men* than women, while I am not aware that sea-
"son or complexion has any influence in the matter. There
"are three marked peculiarities in this disease: 1. Its
"hereditary nature. 2. A tendency to insanity and sui-
"cide. 3. Its manifesting itself as a grave disease only in
"adult life.

"1. Of its hereditary nature. When either or both the
"parents have shown manifestations of the disease, and
"more especially when these manifestations have been of
"a *serious* nature, one or more of the offspring almost in-
"variably suffer from the disease, if they live to adult age.
"But if by any chance these children go through life *with-*
"out it, the thread is broken, and the grandchildren and
"great-grandchildren of the original shakers may rest as-
"sured that they are free from the disease. This, you will
"perceive, differs from the general laws of so-called hered-
"itary diseases, as for instance in phthisis, or syphilis,
"when *one* generation may enjoy entire immunity from
"their dread ravages, and yet in another you find them
"cropping out in all their hideousness. Unstable and
"whimsical as the disease may be in *other* respects, in *this*
"it is firm; it never skips a generation to manifest itself
"in another; once having yielded its claims, it never re-
"gains them. In all the families, or nearly all in which
"the choreic taint exists, the nervous temperament greatly
"preponderates, and in my grandfather's and father's ex-
"perience, which conjointly cover a period of seventy-

“eight years, nervous excitement in a marked degree al-
“most invariably attends upon every disease these people
“may suffer from, although they may not when in *health*
“be over-nervous.

“2. The tendency to insanity, and sometimes that form
“of insanity which leads to suicide, is marked. I know of
“several instances of suicide of people suffering from this
“form of chorea, or who belonged to families in which the
“disease existed. As the disease progresses the mind be-
“comes more or less impaired, in many amounting to in-
“sanity, while in others mind and body both gradually
“fail until death relieves them of their sufferings. At
“present I know of two married men, whose wives are
“living, and who are constantly making love to some young
“lady, not seeming to be aware that there is any impro-
“priety in it. They are suffering from chorea to such an
“extent that they can hardly walk, and would be thought,
“by a stranger, to be intoxicated. They are men of about
“fifty years of age, but never let an opportunity to flirt
“with a girl go past unimproved. The effect is ridiculous
“in the extreme.

“3. Its third peculiarity is its coming on as a grave
“disease, only in adult life. I do not know of a single
“case that has shown any marked signs of chorea before
“the age of thirty or forty years, while those who pass the
“fortieth year *without* symptoms of the disease are seldom
“attacked. It begins as an ordinary chorea might begin,
“by the irregular and spasmodic action of certain muscles,
“as of the face, arms, etc. These movements gradually
“increase, when muscles heretofore unaffected take on the
“spasmodic action, until every muscle in the body becomes
“affected (excepting the involuntary ones), and the poor
“patient presents a spectacle which is anything but pleasing
“to witness. I have never known a recovery, or even an

“amelioration of symptoms in this form of chorea; when
 “once it begins it clings to the bitter end. No treatment
 “seems to be of any avail, and indeed now-a-days its end
 “is so well known to the sufferer and his friends, that med-
 “ical advice is seldom sought. It seems at least to be one
 “of the incurable.

“Dr. Wood, in his work on the practice of medicine,
 “mentions the case of a man, in the Pennsylvania Hospital,
 “suffering from aggravated chorea, which resisted *all*
 “treatment. He finally left the hospital uncured. I
 “strongly suspect that this man belonged to one of the
 “families in which hereditary chorea existed. I know
 “nothing of its pathology. I have drawn your attention
 “to this form of chorea gentlemen, not that I considered
 “it of any great practical importance to you, but merely
 “as a medical curiosity, and as such it may have some
 “interest.”

ZUR CASUISTIK DER CHRONISCHEN HUNTINGTON'SCHEN CHOREA.

Von Prof. Dr. Adolf Strümpell,

• Direktor der medizinischen Klinik in Breslau.

Die chronische *Huntington'sche* Chorea ist eine auch bei uns in Deutschland recht seltene Erkrankung, von der ich selbst im Laufe der Jahre nur etwa ein halbes Dutzend Fälle beobachtet habe. Von einigen früher in *Erlangen* beobachteten Fällen besitze ich leider keine genaueren Aufzeichnungen und kann daher hier nur die Krankengeschichte von *zwei* Fällen mitteilen, die ich in der hiesigen *Breslauer* medizinischen Klinik untersucht habe.

Der *erste* Fall betrifft einen gegenwärtig 46 jährigen *Fleischer* W. F., dessen Leiden im Jahre 1903 begann.

Pat. bemerkte eine auffallende Ungeschicklichkeit seiner *Arme* und *Hände*, sodass er sich öfter kleine Verletzungen mit dem Fleischermesser zuzog. Diese Ungeschicklichkeit war durch abnorme unwillkürliche Bewegungen bedingt, die er selbst nur mit Mühe eine Zeit lang unterdrücken konnte. Erst $1\frac{1}{2}$ Jahre später bemerkte er eine ähnliche Störung auch in den *Beinen*. Der Gang wurde ungeschickter und mühsamer. Pat. musste absichtlich grössere Schritte machen, um sicher zu gehen. Allmählich nahm seine Ungeschicklichkeit mehr und mehr zu. Er stach sich zuweilen beim Essen mit der Gabel, er biss sich öfter auf die Zunge, einmal verletzte er sich aus Versehen mit der Axt am Kopf. Seit ca. 1 Jahre wurde die Sprache undeutlicher und schlechter verständlich. Auch verschluckte er sich häufig beim Schlucken flüssiger Nahrung, sodass heftiger Husten eintrat.—Das Allgemeinbefinden war nicht gestört. Keine Kopfschmerzen. Appetit gut. Harnentleerung ungestört.—In der gesamten Verwandtschaft des Pat. ist seines Wissens *keine ähnliche Erkrankung* bisher vorgekommen.

Status Praesens. Mittelgrosser Mann von ziemlich gutem Ernährungszustand. Liegt Pat. in Bett, so bemerkt man eine ständige Unruhe in den Fingern, weniger in den Zehen, zuweilen auch in den Armmuskeln. Pat. kann nicht lange in derselben Lage verharren. Er wechselt oft die Lage wobei er aber in unkoordinierter Weise ausfahrende Bewegungen mit dem Rumpf, mit den Armen und Beinen macht. *Willkürliche Bewegungen* werden heftig und ausfahrend ausgeführt. Bewegt Pat. die Arme, so verstärken sich die choreatischen Bewegungen in den Beinen und umgekehrt. An den *Augenmuskeln* kein deutliches abnormes Zucken. Wohl treten aber bei Aenderungen der Blickrichtung Zuckungen und abnorme Bewegungen im Kopf auf. Auch in den *Ge-*

sichtsmuskeln für gewöhnlich keine Zuckungen. Nur auffallend häufiger Lidschlag. Sobald aber Pat. die Zunge herausstreckt, treten starke Mitbewegungen in den Gesichtsmuskeln auf, sodass Pat. die merkwürdigsten Grimassen schneidet. Nicht selten greifen diese Bewegungen auf den Kopf und die Arme über. Auch an den *Gaumenbögen* können choreatische Zuckungen bemerkt werden. Die *Sprache* ist verständlich, aber stossweise abgerissen.—Beim *Stehen* beständige Unruhe in den Fingern und den Unterarmen. Der *Gang* ist breit und langspurig. Dabei starke Unruhe in den Armen und Zehen.

An der *Intelligenz* ist keine Störung nachweisbar. *Reflexe* lebhaft, im Ganzen normal. *Pupillenreaktion* ungestört. Sensibilität nach allen Richtungen hin ungestört. Der *Muskeltonus* ist im Ganzen deutlich herabgesetzt. Die inneren Organe alle vollkommen gesund.

Pat. blieb einige Wochen zur Beobachtung in der Klinik. Irgend welche geistige Störungen wurden an ihm nicht beobachtet.

Der *zweite Fall* betrifft einen 41 jährigen Arbeiter J. B., der vom 17. Januar bis zum 5. Februar 1907 in der Klinik beobachtet wurde.

Pat. gibt an, dass die Krankheit, an der er leidet, schon mindestens in *drei Generationen seiner Familie* vorgekommen sei, aber stets nur bei den *männlichen* Familiengliedern. Sein *Grossvater* hat bestimmt an der Krankheit gelitten, ebenso dessen vier Söhne, während dessen Tochter gesund geblieben ist. Pat. hat drei Brüder, die alle an der Krankheit leiden, aber eine gesunde Schwester. Pat. selbst ist verheiratet und hat 5 Kinder, die bis jetzt alle gesund sind (im Alter bis zu 15 Jahren). Bei allen Familiengliedern, die krank geworden sind, ist das Leiden im Alter von ca. 35-40 Jahren zum Ausbruch gekommen.

Dabei hat aber der Grossvater ein Alter von 80 Jahren erreicht, während der Vater schon früh durch einen Sturz verunglückt ist. Eine besondere Gelegenheitsursache zu dem Auftreten der Krankheit ist nicht hervorgetreten. Bei unserem Pat. ist die Krankheit im 38sten Lebensjahre aufgetreten und zwar, wie es scheint, ziemlich gleichzeitig in allen Muskelgebieten. Die unwillkürlichen Zuckungen wurden allmählich immer stärker. Jedoch wechseln Perioden grösserer Unruhe mit Zeiten relativer Ruhe.

Status praesens. Pat. ist ein schwächlig gebauter, etwas blasser Mann. Im ganzen Körper beständige Muskelunruhe. Pat. kann nicht einen Augenblick ruhig im Bett liegen. Er bewegt den Kopf oder runzelt die Stirn, macht eigentümliche, unzweckmässige, regellose Bewegungen mit den Armen oder den Beinen. Soll Pat. mit den Händen einen Gegenstand ergreifen, so fährt er daneben. Auch beim Gehen allgemeine Mitbewegungen in den Armen, im Kopf u. s. w. Im Ganzen ist aber die Muskulatur des Rumpfes weniger von den Zuckungen befallen, als die Muskulatur der Extremitäten. Sehr lebhaft ist die choreatische Unruhe in den Gesichtsmuskeln. *Reflexe* lebhaft, ohne Besonderheiten. *Sensibilität* normal. In den Muskeln der unteren Extremitäten deutliche *Herabsetzung des Muskeltonus*. Intelligenz entschieden etwas herabgesetzt. Pat. ist sehr misstrauisch. Keine gröberen psychischen Störungen. Innere Organe durchweg normal.

Die beiden beschriebenen Fälle sind unzweifelhaft typische Beispiele der echten „*chronischen Huntington'schen Chorea*.“ Dass die Bezeichnung „*hereditäre Chorea*“, wenn auch für die meisten, so doch nicht für *alle* Fälle past, zeigt die erste der mitgeteilten Beobachtungen. Natürlich ist es nicht ausgeschlossen, dass in der Familie bereits Choreafälle vorgekommen sind. Jeden-

fals aber war dem Patienten davon nichts bekannt und es ist natürlich auch durchaus notwendig anzunehmen, dass die Veranlagung zur Chorea auch einmal „spontan“ d. h. ohne unmittelbaren hereditären Einfluss entstehen kann. Wir sehen ja dasselbe bei allen anderen familiären Erkrankungen. Auch bei der „hereditären“ Ataxie, bei der „hereditären“ Formen der Muskelatrophie kommen hier und da scheinbar ganz vereinzelt auftretende Fälle vor. Natürlich werden wir dann aber auch die vereinzelt Erkrankungen nicht auf irgend eine äussere („exogene“) Krankheitsursache, sondern auf eine angeborene („endogene“) krankhafte Veranlagung des Nervensystems zurückführen. Sehr oft sind es freilich dann äussere Einflüsse (Traumen, akute Infektionen u. a.) welche die krankhafte Veranlagung zum Ausbruch kommen lassen. Aber gerade bei der hereditären Chorea scheinen nach allen bisherigen Erfahrungen derartige accessorische Krankheitsursachen nur selten eine Rolle zu spielen. Auch in unseren beiden Fällen entwickelte sich das Leiden scheinbar ganz von selbst.

Eine Eigentümlichkeit der chronischen Chorea, die in einem gewissen Gegensatz zu den übrigen hereditären Nervenkrankheiten steht, ist ihr Auftreten in einem verhältnismässig schon vorgerückten Lebensalter. Während viele Formen der familiären Muskelatrophie hauptsächlich im eigentlichen Kindes und jugendlichen Alter beobachtet und ebenso die hereditäre Ataxie wenigstens meist in jugendlichem Alter, ist die familiäre Chorea bisher noch nie im eigentlichen Kindes- und jugendlichen Alter beobachtet worden. Auch in der Familie unseres zweiten Pat. trat das Leiden bei allen disponierten Familiengliedern erst im Alter von ca. 35-40 Jahren auf. Dies entspricht auch der Mehrzahl der sonstigen Beobachtungen. Nun ist freilich zu bedenken, dass die ersten Anfänge des Leidens sicher

oft lange Zeit unbemerkt bleiben, da die Erscheinungen sich sehr langsam entwickeln und die Kranken sich daher an die leichten Zuckungen so gewöhnen, dass sie dieselben garnicht als etwas Abnormes empfinden. Immerhin ist aber doch das so späte Auftreten der Krankheitssymptome bei einer exquisit hereditären Krankheit etwas Auffallendes. Man könnte die Annahme machen, dass erst eine gewisse *Abnutzung* der krankhaft veranlagten nervösen Centralorgane *durch die Funktion* stattfinden müsse, ehe diese krankhafte Veranlagung zur Geltung kommt. Uebrigens steht dies späte Auftreten der familiären Chorea nicht ganz vereinzelt da. Ich kenne mehrere Fälle von familiärer Muskelatrophie, die sich ebenfalls erst im höheren Lebensalter entwickelt haben.

In Betreff des *Geschlechts* findet man bei den Autoren meist die Angabe, dass Männer und Frauen gleich häufig von der Krankheit befallen werden. Es mag ein Zufall sein, unter meinen eignen Beobachtungen überwiegt das *männliche* Geschlecht ganz erheblich über das weibliche. Dem entsprechend möchte ich auch hervorheben, dass in der Familie meines zweiten Patienten J. B., soweit bekannt, nur die männlichen Familienglieder— diese aber sämtlich!— von dem Leiden befallen wurden, während die weiblichen Familienmitglieder durchweg verschont blieben.

Ueber die *Symptomatologie* des Leidens vermag ich wenig Besonderes zu sagen. Aufgefallen ist mir stets, dass die choreatischen Störungen bei der Huntington'schen Chorea nicht nur in Form einzelner Zuckungen, sondern in besonders charakteristischer und eigentümlicher Weise als *Mitbewegungen* auftreten. Lässt man einen Kranken mit ausgesprochener Huntington'schen Chorea den Mund willkürlich öffnen und die Zunge zeigen, so treten dabei meist im ganzen Gesicht, oft auch im Kopf und in den

Armen die wunderlichsten und bizarrsten Mitbewegungen in Form von Grimassen, Gesticulationen und dergl. auf. Da solche Mitbewegungen alle einfachen Handlungen der Kranken begleiten, so erklärt sich das höchst auffällige Benehmen derselben. Ich habe einen vortrefflichen Universitätslehrer gekannt, über dessen wunderliche Bewegungen und Manieren beim Sprechen, Essen und dergl. man sich lange Zeit lustig machte, bis sich schliesslich herausstellte, dass er an Huntington'scher Chorea litt.

Abgesehen von der motorisch-choreatischen Störung sind eigentlich nur noch die *psychischen Symptome* für die Huntington'schen Chorea charakteristisch. In den meisten der von mir beobachteten Fälle traten die psychischen Symptome nur wenig hervor, oder fehlten sogar, wie in den ersten der beiden mitgeteilten Fälle, ganz. Damit ist natürlich nur gesagt, dass die psychischen Störungen sich oft erst sehr spät entwickeln. Je mehr man Gelegenheit hat, die Kranken genauer zu beobachten, um so früher wird man freilich leichte Störungen der Intelligenz oder der affektiven Sphäre nachweisen können. Der oben von mir kurz erwähnte Fall bei einem Universitätslehrer endete schliesslich mit völliger geistiger Umnachtung.

Sensibilitätsstörungen scheinen niemals aufzutreten. Auch an den *Reflexen* habe ich nichts Besonderes bemerkt. Dagegen möchte ich betonen, dass mir in den beiden oben etwas ausführlicher beschriebenen Fällen eine *Herabsetzung des Muskeltonus* (Hypotonie) auffiel, ähnlich wie sie in schweren Fällen von Chorea Minor nicht selten beobachtet wird. Die Ursache dieser Erscheinung ist noch nicht völlig klar. Es scheint sich um eine Verstärkung der hemmenden cerebralen Einflüsse zu handeln—im Gegensatz zum Fortfall der Hemmungen bei cerebralen *Lähmungen*.

Ueber das eigentliche *Wesen der choreatischen Symp-*

tome können wir einstweilen noch kaum eine Vermutung aussprechen. Größere anatomische Veränderungen der Centralorgane sind sicher nicht vorhanden und auch von vornherein garnicht zu erwarten. Es kann sich nur um Störungen in der funktionellen Organisation der normalen motorischen Innervation handeln. Für den normalen motorischen Betrieb unseres Körpers ist die ruhige Fixation und die jederzeit mögliche Unterdrückung der Innervationen ebenso wichtig, wie der richtige motorische Antrieb. Diese allgemeine Regulierung der gesamten Muskelinnervation ist offenbar bei der Chorea chronica in völlige Unordnung geraten. Welche Vorgänge aber dieser allgemeinen Auflösung aller Ordnung zu Grunde liegen, ist einstweilen noch völlig unbekannt.

LA NATURE DE LA LÉSION HISTOLOGIQUE DE LA CHORÉE DE HUNTINGTON.

Par M.M. les Docteurs

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La chorée de Huntington, chorée héréditaire, occupe actuellement dans le cadre des chorées chroniques progressives une place bien définie basée sur des caractères cliniques précis et sur des lésions anatomiques suffisamment constantes. Nous avons encore retrouvé ces dernières dans l'observation qui sert de point de départ à cette courte note.

Mais si la lésion histologique (infiltration de grains prédominant dans l'espace péricellulaire avec altérations plus ou moins marquées des cellules pyramidales) est

aujourd'hui admise sans conteste en tant qu'altération évidente au microscope, sa nature intime reste encore sujette à controverse. Nous allons même voir qu'elle n'est pas absolument spéciale à la chorée de Huntington. Celle-ci ne serait plus qu'un cas particulier, d'ailleurs nettement défini par son étiologie et son évolution clinique, dans un groupement synthétique d'affections caractérisées par des mouvements anormaux dont la cause première serait une irritation histologique des cellules pyramidales ou des faisceaux moteurs qui en émanent.

Voici d'abord notre nouvelle observation :

Observation.

J. B. Let., 61 ans, ferblantier, entré à l'hôpital en mai 1902. Père mort à 69 ans d'affection cardiaque : il était devenu choréique à 45 ans et présentait des mouvements beaucoup plus marqués que ceux de son fils. Une cousine paternelle présente actuellement la même affection choréique—Pas d'autres maladies nerveuses dans la famille : il a perdu un frère à 35 ans qui n'était pas choréique et une soeur de 46 ans est actuellement indemne de la maladie. Il a 5 enfants non choréiques : il a perdu une fille qui était hystérique et cataleptique.

Alcoolisme et absinthisme avéré.—Pas de syphilis.

Il ne peut préciser le début : il bougeait déjà depuis plusieurs années, mais sans avoir cessé son travail, lorsqu'il y a 5 ans, il se fit une aggravation subite. Depuis 3 ans, il ne peut plus du tout travailler.

Mouvements incoordonnés et très étendus au niveau des membres supérieurs qui s'agitent sans arrêt. Ils sont également incessants au niveau des membres inférieurs et surtout des pieds. Il peut cependant marcher, mais tombe souvent : aussi ne sort il plus seul dans la rue. Mouvements du tronc, du cou et de la tête. Rien du côté de la face, de la langue ni des yeux. La force est bien conservée

sauf au niveau de la main gauche qui serre mal. La volonté atténue les mouvements sans les arrêter complètement. Sensibilité intacte. Reflexes rotuliens exagérés. Aucune lésion viscérale.

Comme troubles psychiques, on note de la gêne de la parole. Amnésie nette: il ne sait plus son âge. De plus un peu d'idées délirantes sur sa situation: il se dit riche, ayant gagné beaucoup d'argent dans son métier, alors qu'en réalité il est sans ressources.

A la fin de décembre 1902 il présenta un état aigü pulmonaire avec crises de suffocation, tachycardie, albuminurie et élévation de température jusqu'à 39°5 qui dura huit jours et fut marqué par une recrudescence des mouvements: il devint incapable de se rendre le moindre service, de manger, de s'habiller, etc.

A partir de ce moment il se mit à décliner rapidement, cessa de s'alimenter, maigrit beaucoup.

Le 16 février il prit une ulcération sur la cornée droite et trois jours après l'oeil avait subi une véritable fonte. Le 1^{er} mars, on note une ulcération semblable sur la cornée gauche avec cécité totale. Le 18 mars ulcération sur la face dorsale de la main gauche. A ce moment on note que les mouvements qui s'étaient beaucoup atténués ont complètement cessé depuis 3 ou 4 jours, sauf au niveau de la tête. Somnolence continue qui se termine par le coma et la mort le 24 mars 1903.

Autopsie 24 heures après la mort:

La dure-mère est épaisse et adhérente à la calotte crânienne: pas d'adhérences entre la dure-mère et la pie-mère. Le pôle antérieur du cerveau est recouvert d'un caillot hémorrhagique qui est surtout sus-arachôïdien. Oedème marqué dans les mailles de la pie-mère.

L'encéphale est petit et ne pèse dans sa totalité que 1030 grammes.

La pie-mère se détache très facilement à gauche: il y a quelques petites adhérences du côté droit. Un peu au-dessous du pli courbe à droite on trouve dans la pie-mère une petite tumeur grosse comme une noisette qui paraît fibreuse et a seulement creusé une petite dépression dans l'écorce.

Les circonvolutions sont normales, un peu grêles, sans grosses lésions: on ne trouve rien sur les coupes du cerveau, du cervelet, de la protubérance et du bulbe.

Pas de lésions macroscopiques de la moëlle.

Du côté des viscères on trouve de la symphise pleurale des deux côtés. Pas de lésions des poumons d'aucune sorte, en dehors d'un peu de bronchite purulente.

Le coeur est très petit, ne pèse que 180 gr., mais, en dehors d'un peu d'athérome de la base des valvules aortiques et de l'aorte, ne présente aucune lésion.

Foie petit également, pesant 1130 gr., un peu congestionné, sans lésions macroscopiques. Grosse vésicule remplie de calculs et de liquide d'aspect puriforme.

Les reins ne pèsent ensemble que 200 gr., se décortiquent bien, n'offrent à la coupe aucune lésion macroscopique, sauf une cicatrice peu étendue dans la substance corticale de l'un d'eux.

EXAMEN HISTOLOGIQUE: *Zone motrice droite.* (Inclusion à la celloïdine; coloration par la méthode de Nissl). — Dès l'examen à un faible grossissement, on voit la zone des cellules de la couche corticale de la circonvolution parsemée de petits grains vigoureusement colorés en bleu. Ces grains bleus, homogènes, apparaissent, à ce grossissement, comme sur un plan plus superficiel que les cellules nerveuses. Ces grains sont rares dans la couche moléculaire de la surface de la circonvolution, ils sont exceptionnels dans l'axe blanc de la circonvolution.

A un fort grossissement, ces *grains bleus* offrent par

l'homogénéité de leur chromatine, l'intensité de leur coloration, leur plus petit volume des caractères qui les différencient immédiatement des grains de la névroglie. Dans la zone des cellules nerveuses ils sont souvent groupés par 3 ou 4 autour d'une même cellule pyramidale. Tantôt sur un plan plus profond, tantôt sur un plan plus superficiel, tantôt accumulés à la base ou sur un côté de la cellule nerveuse, ces grains bleus peuvent paraître comme pénétrer cette cellule. C'est-à-dire que, au voisinage d'une cellule pyramidale, ces grains bleus sont comme au centre d'un halo clair qui, pour une certaine mise-au-point, semble creuser le protoplasma de la cellule nerveuse. D'ailleurs, on peut remarquer que toujours le grain bleu reste à la périphérie de la cellule nerveuse ou de l'un de ses bras, son halo clair encoche le protoplasma plus ou moins profondément, mais il n'y disparaît pas. Quand le grain paraît incorporé au protoplasma de la cellule nerveuse, il est aisé de voir qu'il se trouve sur un plan ou plus superficiel ou plus profond.

Si l'on considère les *cellules nerveuses* de la corticalité, on remarque qu'aucune ne présente sa substance chromatophile à l'état normal; sur toutes il y a au moins de la chromatolyse diffuse. De plus, fait constant, les cellules pyramidales sont toutes déformées; elles sont devenues le plus souvent étoilées, creusées de dépression sur leurs trois faces, leurs bras sont devenus grêles, souvent tortueux, leur noyau le plus souvent ne serait plus visible, s'il n'était marqué par son nucléole. Les grandes pyramidales elles-mêmes sont le plus souvent très pâles et difficiles à bien voir. En somme, leur principale et constante altération, immédiatement appréciable, est constituée par ce changement de forme qui tourmente et leur corps protoplasmique et leurs prolongements. Enfin dans les zones où les grains bleus sont plus abondants, il est de suite

patent que ces altérations des cellules nerveuses sont plus marquées.

Zône motrice gauche.—Mêmes lésions et de même intensité que sur les coupes du fragment de la zone motrice droite.

Noyaux gris centraux.—L'infiltration des petits grains bleus disséminés d'une façon diffuse au travers des cellules nerveuses est encore plus intense que sur les coupes de la corticalité cérébrale. De plus là, d'une façon beaucoup plus marquée, des artérioles présentent dans leur gaine périvasculaire un manchon de petits grains bleus; à côté de celles qui offrent un manchon absolument continu, il y a quelques artérioles qui en sont complètement exemptes.

Les cellules nerveuses, de forme normalement plus irrégulière dans les noyaux centraux, présentent le même type de chromatolyse, le même envahissement de leur logette cellulaire par les grains bleus.

Dans les bandes de substance blanche qui interrompent et traversent la nappe des cellules nerveuses de ces noyaux, les grains bleus se présentent avec leurs mêmes caractères morphologiques, mais ils se disposent en file, comme en lignes parallèles à la direction des fibres nerveuses.

Nous avons pu constater sur des coupes des noyaux gris centraux colorées au carmin pendant 48 heures, qu'on ne pourrait avoir idée de cette lésion, que par cette coloration elle échapperait certainement.

Dans un mémoire qui remonte aujourd'hui à dix ans⁽¹⁾ étudiant deux cas de chorée héréditaire, nous avons émis une hypothèse pour expliquer la lésion de Greppin, que nous avons retrouvée constante dans tous les cas que nous avons eu à examiner. Nous avons pensé qu'il s'agissait d'une véritable malformation héréditaire de la

(1) Lannois et Paviot.—Deux cas de chorée héréditaire avec autopsies.—*Revue de Médecine*, 10 Mars, 1898.

névroglie surtout de la substance corticale, et nous expliquions l'apparition plus ou moins tardive des manifestations choréiques dans cette maladie familiale par l'accentuation progressive de la malformation tératologique. Du reste, ajoutons-nous, on peut admettre avec Dana que les cellules pyramidales elles-mêmes ont une tare originelle héréditaire . . . et cette hypothèse répond par avance aux cas où on pourrait observer des lésions analogues à celles qui ont été décrites par Greppin, par Dana, par Clarke, etc., et par nous, sans qu'il se soit produit cliniquement de mouvements choréiformes.

Depuis ce mémoire, non seulement nous avons pu vérifier la présence des granulations dans l'écorce et les noyaux gris centraux d'un autre cas de chorée héréditaire,⁽²⁾ mais nous l'avons rencontrée dans d'autres affections similaires ou très voisines comme symptomatologie. Récemment nous avons pu constater dans un cas de chorée symptomatique liée, selon toutes probabilités, à l'alcoolisme, une infiltration presque identique de petits grains bleus dans l'écorce et le centre blanc des circonvolutions. Il en a été de même dans un cas, que l'un de nous va publier, de myoclonie.

Il semble bien à l'heure actuelle, comme l'ont exprimé Murri (de Bologne), Roncoroni, Mannini, que les chorées, les myoclonies et certains tics vrais ont une parenté non seulement clinique, mais histologique. Nous ajouterions certaines athétoses doubles, car nous avons pu observer un cas de grande athétose, où les lésions étaient réduites à une méningo-encéphalite de la région rolandique. Le trait-d'union entre ces diverses manifestations symptomatiques serait constitué par une véritable encéphalite déterminant l'altération progressive plus irri-

(2) Lannois, Paviot et Mouisset.—Contribut. à l'anatomie pathologique de la chorée d'Huntington.—*Revue Neurol.*, 1901.

tative que destructive de la couche corticale cérébrale et surtout de la zone rolandique. Le trait-d'union entre ces diverses manifestations symptomatiques serait constitué par une véritable encéphalite déterminant l'altération progressive plus irritative que destructive de la couche corticale cérébrale et surtout de la zone rolandique. Cette notion que, pour notre part, nous avons plusieurs fois défendue a été nettement exprimée par Murri: "lorsque une lésion de cette zone, dit-il, ne produit ni clonie, ni chorée c'est qu'elle en diminue l'excitabilité et produit la parésie ou la paralysie; clonie et chorée demandent, pour être réalisées, l'excitabilité."

Nous pensons que rien n'empêche de faire rentrer la chorée de Huntington, pour héréditaire qu'elle soit, dans la même loi. Ce qui est héréditaire dans la maladie, c'est le terrain favorable à l'apparition d'affections du système nerveux. Que chez un membre de la famille offrant dans ses ascendants ou ses collatéraux des cas de chorée de Huntington survienne une infection ou une intoxication, sa prédisposition peut entrer en jeu, son système nerveux et notamment son écorce cérébrale appelleront, pour ainsi dire, la localisation infectieuse ou toxique. Les exemples d'hérédité de terrain et d'hérédité similaire seraient faciles à trouver pour les maladies du système nerveux.

HISTORICAL NOTE ON HEREDITARY CHOREA.

By William Osler, M.D., F.R.S.,

of Oxford, England.

While working at the subject of Sydenham's chorea, in 1886-87, I became interested in other forms, and my attention was directed to the hereditary variety described as prevalent in the eastern part of Long Island. I wrote to Dr. George Huntington, and in a letter dated La Grangeville, New York, June 9, 1886, he gave me a few additional facts and referred me to Dr. Osborn, of Easthampton. From the latter I obtained a good deal of information about the distribution of the disease, and I had arranged with him to visit the place, but he wrote in July, 1887, that the subjects were so sensitive that he did not think I could get access to them if I came. In 1898 Dr. Osborn again sent me notes of the cases.

It may be interesting to follow briefly in the literature the original description of the three chief sub-divisions of the diseases we now speak of under the name chorea.

The name came from the dancing mania which had prevailed extensively throughout Europe in the middle ages—an epidemic of a remarkable character in which hundreds, even thousands of persons became possessed with a common delusion and danced through the country like raving maniacs. The malady was believed to be due to the work of demons, but, as Hecker says, Paracelsus, that "mighty, but as yet scarcely comprehended reformer of medicine, whose aim it was to withdraw diseases from the pale of miraculous interpositions and saintly influence and explain their causes upon principles deduced from his knowledge of the human frame," would not admit for a

moment that the saints had any power to inflict disease, and he protested that diseases should not be named for them. The dancing mania had been popularly known as St. John's dance, St. Vitus's dance. He gave the name of Chorea to St. Vitus' dance and spoke of three forms: *C. imaginativa* by which he described the original dancing mania; second, that which arose from sensual desires, *C. lascivia*; and thirdly, that which arose from corporeal causes, *C. naturalis*. An excellent account is given of this dancing mania in Hecker's *Epidemics of the Middle Ages*.*

With the common everyday type of the disease in children one of the greatest names in medicine is associated. It was in his *Epistolæ Responsoræ*, 1680, that Thomas Sydenham described the Chorea Minor. It was not a very happy name to have given to the disease. He may, of course, have got the original description from Paracelsus; possibly from one of the earliest references to it which I find in English literature, viz., Burton's "Anatomy of Melancholy." Sydenham's account has always been regarded as admirable and it is worth quoting: "Chorea Sancti Viti is a sort of Convulsion, which chiefly invades Boys and Girls from ten Years of Age to Puberty: First, it shews it self by a certain Lameness, or rather Instability of one of the Legs, which the Patient drags after him like a fool; afterward it appears in the hand of the same side; which he that is affected with this Disease, can by no means keep in the same Posture for one Moment, if it be brought to the Breast or any other part, but it will be distorted to another Position or Place by a certain Convulsion, let the Patient do what he can. If a Cup of Drink be put into his Hand, he represents a thousand Gestures, like Juglers, before he brings it to his Mouth;

*Sydenham Society, 1844.

for whereas he cannot carry it to his Mouth in a Right-line, his hand being drawn hither and thither by the Convulsion, he turns it often about for some time, till at length happily reaching his Lips, he flings it suddenly into his Mouth, and drinks it greedily, as if the poor Wretch designed only to make sport." (Pechy's translation.)

The description of Hereditary Chorea, or as it is perhaps better called, the Chronic Progressive, is an interesting chapter in American medicine. As is so often the case, no one man deserves all the credit, and I know Dr. Huntington is quite ready to acknowledge the good work of those who preceded him. In 1842 Dr. C. O. Waters, of Franklin, New York, recognized the disease, and reference is made to it in the first edition of Dunglison's "Practice of Medicine." He recognized the onset in adult life, the hereditary characters and the incurable nature. In 1863 Dr. Irving W. Lyon wrote an article in the *American Medical Times*, December 19th, on Chronic Hereditary Chorea, in which he recognized practically all the features of the disease. Within the past year I have had correspondence with his son, my friend and old pupil, Dr. Irving P. Lyon, of Buffalo, who has presented very strongly his father's claims to be recognized as the original describer of the disease. I have, unfortunately, mislaid the copy which he sent me of his father's paper and his own letters, but I have asked him to append a short memorandum, as I know Dr. Huntington and others would like to have full justice done to his memory.*

In the history of medicine there are few instances in which a disease has been more accurately, more graphically, or more briefly described than that in which Dr. Huntington calls attention to an *Hereditary Chorea* which prevailed at the eastern end of Long Island, where

*Full credit is given in the sketch of Lyon (p. 147) and in the Bibliography (p. 153).—Ed.

both his father and grandfather had practised. The paper appeared in the *Medical and Surgical Reporter*, April 13, 1872, as an appendix to an everyday sort of paper on Chorea Minor. In my monograph on Chorea, 1889, his account was reproduced in full, I believe, for the first time, and it is very appropriate to reprint it here. (See page 95). The chief features of the disease are very clearly sketched—the hereditary nature, the progressive dementia and the onset in adult life.

A CONTRIBUTION TO THE HISTORY OF HUNTINGTON'S CHOREA.—A PRELIM- INARY REPORT.

By Smith Ely Jelliffe, M.D., Ph.D.,

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During the past five or six years I have been trying to collect data concerning the hereditary origin of some cases of Huntington's chorea in the United States, meaning by this a study of some of the early families in which the disease has appeared, and the tracing of its spread throughout the United States through the progeny of some of these affected stocks. The work has been of considerable interest, although the amount of time which such a study has consumed is out of all proportion to the results obtained. This has been so for a number of reasons which it is not here necessary to record in detail, but may be readily appreciated by all who have attempted any work of this particular character.

Such a study involves a large amount of genealogical research through imperfect records, and a more or less futile search of the histories of many early colonial settle-

ments, particularly with reference to emigration of families from one state or county to another, with an interminable correspondence with members of afflicted families and with physicians who have reported cases, or who have had patients suffering from the disease under their care.

I desire to express my sincere thanks to these physicians and laymen who have so generously answered my letters. I cannot mention them here by name as they are so many, but my present communication would have been impossible without their co-operation.

The fragmentary character of many genealogical trees, the unwritten colonial histories, the reticence of families to reveal their family skeletons, and the lack of knowledge on the part of physicians of the actual names of the forbears of their afflicted families have been among the difficulties encountered.

This has made it impossible to get all of the evidence in a satisfactory state for final presentation and I have consented to give for the present Huntington number only a preliminary statement of some of the facts brought out, because of the urgent request of Dr. Browning, and further because such a statement may make it easier for myself or for others to fill out the picture here sketched in its bleakest and most fragmentary outlines. I feel constrained in this place to say that the facts can be alluded to here only; the complete evidence must wait for more mature summaries.

At the outset it may be well to say that the present communication concerns itself solely with a review of the early families, from whom our original descriptions came. The foreign cases who have come into the country, in great numbers, and have been a very difficult conflicting feature in this study, are neglected entirely.

Wharton Sinkler says in a paper on Huntington's Chorea, page 71¹: "I cannot help thinking that the cases referred to by Dr. Waters are in all probability the same which Huntington has reported. The latter says that he, his father and grandfather have known families in Long Island in which this form of chorea has lasted for generations. Dr. Waters speaks of the disease being common in the southeastern portion of the State of New York, and this would correspond with the locality of Long Island, and the character of the cases is remarkably like those of Huntington."

"Another paper on hereditary chorea appeared in 1863, and this, strangely enough, seems to have escaped the attention of Dr. Huntington and the writers on the subject who have followed him. These cases are from New York, and may also be from Long Island, but as no names or addresses are given it is impossible to trace any connection between them."

Dr. Sinkler here refers to the paper by Dr. Irving W. Lyon, a House Surgeon in Bellevue Hospital in 1863. Two of Lyon's cases came from Westchester County, New York, one from Greenwich, Connecticut, so that neither of them were from Long Island, directly, as Sinkler suggested, nor were Water's cases Long Island cases either, so that although the idea was in Sinkler's mind, what evidence there was was negative rather than positive. Sinkler² reports three other cases, and on page 285 speaks of the possible spread of the disease from an original source.

But the idea of the spread was not Sinkler's alone, for we find that King,³ in a foot note to an article on Huntington's Chorea says:

¹*Journal of Nervous and Mental Disease*, Feb., 1889.

²*Medical Record*, March 12, 1892.

³*Medical News*, 1889, July 13, p. 41.

"My father's native town, Easthampton, Long Island, was settled as early as 1649, and choreic families have lived there for generations. Previous to 1800 there were several emigrations from the town to eastern and northern localities, one of these being to the western shores of Lake Champlain, near Plattsburgh, another to the Mohawk Valley in Oneida County, another to Delaware County, and a fourth one to some place in New York. It is interesting to note that Dr. Water's observations were made upon patients living in Franklin, Delaware County, two of the cases given by him were in New York State, town and county not given—Huntington's cases were from Eastern Long Island, and Sinkler's first patient was born in New York State. The father of my second patient came from Vermont, town unknown, and the parents of my present patient came from Oneida County, New York. One of Lyon's cases was in Connecticut, only a short distance by water from Long Island, and Sinkler's second case was in Wyoming, Pa., only seventy-five miles from Delaware County, N. Y. My first choreic patients were of Welsh descent, the great grandfather (who afterwards had the disease) coming from Wales and settling in Vermont. He afterwards moved to Plattsburgh, New York, and still later came to Western New York. *Possibly if we were in possession of the complete ancestral history of our patients we could trace all the cases of the disease in this country except my first case, to one center of origin in Easthampton, Long Island, or is this coincidence merely accidental?*"¹

Further Sinkler remarks in his paper in the *Medical Record*, vol. 41, page 285: "King refers to a circumstance to which I also alluded in my previous paper, namely, the probability that many, if not most, of the

¹ Italics mine, S. E. J.

cases reported in this country are of common ancestry. Easthampton, Long Island, according to King, was settled in 1649, and choreic families have lived there ever since. Several emigrations have taken place from there; one of them to Delaware County, New York, and it was from Franklin, Delaware County that Dr. Water's original observations were made. Charles B. Gorman in his thesis, quoted by Dunglison, speaks of the affection existing in Luzerne County, Pa., which is less than 75 miles from Delaware County, New York. One of my cases was from Wyoming County, Pa., which adjoins Luzerne."

This is the thesis which has attracted my attention, and the evidence which I here present, although as yet but fragmentary, I hope may be a precursor of a fuller report, and may serve as a stimulus to others to trace back the genealogy of the family trees, so that some day a complete history of this disease may be presented from this standpoint. Such a study would prove of invaluable assistance as offering evidence bearing on the question of heredity some remarks concerning which I trust may be justified by even the scanty outline which I here present.

As will be noted in the Biographical Sketches offered by Dr. Browning, Dr. Waters was born in Franklin, New York. His medical activities were not carried out at Franklin, and it is probable, as Dr. Browning has pointed out, that Waters referred to Westchester County as the southeastern part of the State. Further it may be pointed out, in addition to the philological argument that Browning adduces, that Water's parents came from Hebron, Conn., and his trips from Franklin to Hebron, through North Salem and Bedford may have led him in this region where much Huntington chorea existed in Lyon's day, and exists even more at the present, from

this original "Bedford" source. This group, following Browning we shall call the group II, Bedford Group.

Irving W. Lyon was born at Bedford, Westchester County, N. Y., in 1840. He lived there until he moved to New York City as a student, and in 1863 was an interne at Bellevue. The cases which he described were seen in Bedford, Westchester County, in Greenwich, Conn., and within a radius of twenty miles from Bedford. His cases are, therefore, of the II Group as well.

Dr. George Huntington, his father and grandfather lived in and about Easthampton, Long Island, and his cases were all in old Long Island families. Group I, Long Island or Easthampton Group.

There is little ground for doubt that Gorman's group was the III or Wyoming Group, and is the group in which one of Sinkler's families is to be arranged. Another would appear to be arranged as coming from the Bedford Group.

Group I.—Easthampton Group—Huntington.

It is not established that this group is the earliest by any means, but it is a fair assumption, from what is known of the history of Easthampton, that after its settlement from Connecticut in 1649 and natural internal growth it suffered more from emigration than it did from immigration.

The original sources of the disease we shall never know. It can only be stated that the afflicted families who lived in Easthampton in the days of the father and grandfather of Huntington were among the earliest of the settlers of the Island. Among the early choeric settlers of the Island we find the names of D....n, H....s, P...e, S...e, O.....e, V.. S..y and C....t. These have all suffered from intermarriages, and although tradition has it that the D....n family is

the original family it is highly improbable that this can be proven.

In Howell's History of Southampton, we read that emigrations took place as early as 1665, to Elizabethtown, N. J., and from this time, for fifty or sixty years members of certain families left Southampton to go to this and adjacent New Jersey towns, Cohansey, Salem County. We think it can be shown that a large group of patients, among whom Collins' case is to be reckoned, sprang from this group. The chief settlers here came from the C...g, P...e, and S...e families into both of which, by intermarriage with the D...n, and H...e families the disease was introduced. One of the New Jersey families is to be definitely traced to Group I, or the Easthampton Group.

In 1800 another emigration went to Montrose, Pennsylvania. This is in Susquehanna County, just ten to fifteen miles north of the Wyoming County line, which county is about twenty miles wide, and immediately north of Luzerne County in which Gorman's cases were found. The original settlers of Montrose included among the names of P...e, H...s, by marriage, which were among the afflicted families of the Easthampton Group. Thus I think it is highly probable that the Gorman cases, or Group III, are directly derived from the Long Island Group.

Sinkler's cases are in part derived from this Wyoming or Gorman Group; while in part descendants of the Bedford Group.

One of the most interesting phases of this study has been to trace the origins of the Bedford Group. I am persuaded, more from general facts than from definite links in my chain of evidence, that these Westchester families were old Connecticut families, and the probab-

ities are that both the Long Island Group, and the Bedford Group sprang from an original Connecticut source. Such a source still shows in numbers of cases scattered throughout the State of Connecticut, the names of some thirty of which have been communicated to me by Dr. A. R. Diefendorf of Yale University.

Quite recently I have met with a group of cases from Vermont, and about Plattsburgh, New York. The M...n family is the present heritor, but coming from the mother's side H...s in name. These are directly traceable to intermarriage with the Easthampton stock that emigrated to Plattsburgh in the early part of the eighteenth century.

At least two of Hamilton's recently reported cases (*Am. J. Insanity*, Jan., 1908), belong to the Easthampton stock.

Dr. Marion's cases in New England and others there still unreported seem to me, so far as I can ascertain, a distinctly separate stock. This introduces a Group IV, or the Massachusetts Group, concerning which I shall speak at a later time.

Riggs' cases are undoubtedly from the Easthampton, or common Connecticut stocks. His earliest ancestor is a S...h, a name unfortunately so widespread in the United States as to baffle the most assiduous student of family trees.

Concerning the hereditary features of Huntington's chorea certain facts are of extreme importance. In the first place it is far from clear that other processes are not capable of causing conditions so closely resembling this disorder as to be unrecognizable by our present clinical differentiations. Certain cases of katatonic præcox enter into the asylum records as Huntington's chorea. These should be rejected. The cases showing no heredity what-

ever are in need of closer scrutiny. Family scandals, skeletons, etc., are difficult to probe into, but in at least three cases apparently non-hereditary choreas were in reality hereditary, for the non-choreic fathers that should have been, were caring for the descendants of choreic fathers in actuality. This is only one of the factors of experience that contribute to the misreading of statistics. All in all my figures seem to show a very close approximation to the results of Mendelian crossing as more recently brought out by Farabee, Drinkwater, Bateson and Punnett (*Proc. Roy. Soc. Med.*, Epidemiological Section, Feb. 28, 1908), but until I have been able to fill out more of my family trees I am unwilling to present any conclusions.

In closing this short note, I again call attention to its preliminary character, and offer it as an invitation for those who may read it, and who have had cases of this disease under their observation, to send me as full a family tree of the afflicted families as possible. One family tree often ties a dozen heretofore unrelated families together.

A FAMILY IN WHICH THE CHOREIC STRAIN MAY BE TRACED BACK TO COLONIAL CONNECTICUT.

"An heirloom from generations away back in the dim past."—*Huntington*.

By Frederick Tilney, A.B., M.D.,

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at the Long Island College Hospital, Etc., of Brooklyn, N. Y.

Concerning the Connecticut and apparently older side of the Bedford Group, common tradition in that region attributes the derivation of the choreic strain entirely to

the "P" family. The purpose of this short note is primarily to carry back an established choreic history further than has hitherto been done, and, for the convenience of future investigators, casually to show that this heredity came into the "P" family in 1777, by marriage, from the Welles or Wells family.

No attempt is here made to indicate the collateral occurrence of this disorder, as this would include too vast a number of cases and only serve to confuse the plain line of descent. Whence or when it came into the Welles family is not stated; but that it came therefrom into the "P" and subsequent families is very definitely asserted and appears to be circumstantially verified. The tracing has been possible because of the fact that members of the family have lived continuously at the same place (Stratford, Conn.) since its first settlement, and have thus preserved much family history. Cases known to have been affected are indicated on the chart by an appended circle.

The patient whose case serves as the starting point of this tracing was first seen by the writer during the winter of 1908 in the Neurological Department of the Polhemus Memorial Clinic. She had already been suffering from her affliction since her thirty-eighth year, being at present forty-six years of age.

During this interval of eight years, her symptoms have been steadily on the increase. The choreic movements which she now presents are so severe as to render all voluntary acts, including locomotion, most uncertain. Her memory has failed to a marked degree; her mental competence is not what it was.

Her family tree clearly indicates that the disease was transmitted to her from her father's side. It is to be noted, however, that her father was not a victim of the

Gov. Thomas Welles.
1598-1660.

John Welles.
1621-1659.

Miss. Hunt.
England.

1647.

Elizabeth P

Robert Welles
1661-1714.

Joseph Welles.
1680----

Joseph Welles.
1720-1738.

Another daughter.

Gift

1739.

Martha Welles
1757-1798.

Martha Curtis.

1777.

Charles P.
1790-1800.

Job P.-K.
1753-1797.

Lewis P.-
killed in Civil
War act. 26.

Monnah Davis.

Harriot P.-
only child.

Sarah R.

malady, and that the only other representatives of his generation (two brothers) also escaped. In this connection it should be mentioned that the father was killed in the Civil War at the early age of twenty-five years. With this in view he may well be considered as having been potential in the transmission of the disease without having suffered from it himself.

The patient's paternal grandfather, on the other hand, presented a perfectly typical case. While this strain is said to have come from his mother, Martha Welles, the latter appears to have escaped; but here also, account must be taken of the fact that this Martha Welles died when still young (being in her forty-first year at the time of death). Two of her sisters (one born before 1750 and the other after that year), were affected. Inasmuch as Martha Welles transmitted the disease to one of her sons, there were therefore three representatives of her generation who carried the choreic strain. This shows that it must have come from a common stock farther back, *i. e.*, from one or other parent, and tradition says from the paternal or Welles side.

In any case, this traces the heredity well back into the first half of that century, giving a known line of descent of nearly two hundred years.

Genealogical References.

History of the Welles Family, By Albert Welles, N. Y., 1876.

History of Stratford, Conn., By Rev. Sam'l Orcutt, N. Y., 1886.

MENTAL SYMPTOMS OF HUNTINGTON'S CHOREA.

By A. R. Diefendorf, A.B., M.D.,

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Conn.

The mental symptoms of Huntington's chorea have always been recognized as one of the three essential elements in the disease picture. From the first it has been quite generally recognized that these symptoms are chiefly those of a gradually progressive deterioration ultimately leading to absolute dementia. The basis for my discussion of these symptoms is the personal observation of fourteen cases, and very full histories given by physicians and relatives of fourteen others. I have collected altogether sixty-five unpublished cases of Huntington's chorea, most of them from Connecticut, but in many of these the history is altogether too meagre to permit of very accurate deduction as to the mental symptoms.

The mental symptoms of Huntington's chorea may be divided into three groups: (*a*) the first and most important being gradually progressive dementia; (*b*) cases with crazy symptoms, and (*c*) cases with symptoms of congenitally defective constitution. The second and third groups, those with the crazy symptoms and with congenitally defective constitutions, are in most cases but episodes in a psychosis which sooner or later ends in a progressive dementia.

Congenitally Defective Constitutions.—These symptoms consist chiefly of a history of excessive "nervousness, irritability and excitability" which appear in childhood and exist throughout puberty and early manhood, usually becoming more pronounced as the individual approaches

middle age, the period when choreic manifestations most often develop. In only two of my twenty-eight cases were there evidences of congenital defect so pronounced as to be classed as imbecility. In one case the imbecility was of a moderate degree, consisting in a certain dullness in school and inability to acquire knowledge readily, some seclusiveness and egotism. In the other case there was marked imbecility, but in this case the child was born a choreic and has continued one up to his present age of eighteen. The congenital mental defect in this case has thus far remained unchanged. Stephens also reports two cases in which the chorea developed in infancy. Symptoms of congenitally defective constitution occurred in eight (28%) of the twenty-eight cases. There should probably be classed in this group another unusual case, that of epilepsy which developed at seventeen and continued unaccompanied by mental symptoms until fifty-three, at which time pronounced epileptic befogged states, rapidly progressive dementia and choreic movements developed coincidentally, the latter of which, however, have recurred only in connection with marked epileptic phenomena. This case is here given in detail for the purpose of record.

E. H., aged 56. Onset epilepsy at seventeen, attacks two to ten monthly. A successful housewife and a mother of eight children. No mental deterioration until fifty-two when there appeared slight simple deterioration with loss of memory, paralysis of thought, etc. Choreic manifestations developed at fifty-three, thence very rapid deterioration until fifty-four when profoundly demented with almost no memory, not even knowing her own name, extreme paralysis of thought, total apathy, no insight, complete disorientation, marked blunting of attention, lack of judgment and great anergy. The

choreic symptoms first occurred during severe and prolonged post-epileptic befogged states, entirely disappearing in the interval, and thus far (fifty-six years) they recur only during such epileptic states.

There is one case reported by Jolly in which chorea and epilepsy developed coincidently at nine years, and two cases by Hoffmann in which epilepsy antedated chorea by several years, also one case by Remak in which epilepsy existed from twenty-three to thirty-one, while chorea did not develop until forty.

Simple Progressive Dementia.—Of my twenty-eight cases, there are eleven showing simple progressive dementia. The usual order of the development of the symptoms was, first, the appearance of an *increasing emotional irritability*, in passionate outbursts, abuse and sometimes even violence and destruction of articles in the environment. Very often the evidences of excessive irritability occurred only in distinct periods, which varied from a few hours duration to two or three weeks. Sometimes associated with the irritability there developed some suspiciousness, particularly in connection with the conduct of those in intimate association with the patient, more especially the husband or wife. Either in connection with the periods of increased irritability or independently of them, there also sometimes developed transitory periods of *despondency*, lasting from a few hours to a few days. The despondency was usually simple in nature, consisting most often of a gloominess and ennui. During this period patients very often threatened and sometimes attempted suicide. Strangely enough the suicidal threats and attempts did not originate from insight into their disease and dread of its development.

In addition to these periods of increased irritability, there usually occurred at the same time a gradually

increasing *emotional deterioration*. This was shown in a variety of ways, but was most often evident in connection with the patient's interests; his work, his home and his associates. During this early period, some patients became intemperate, others immoral. A number of my men patients gave up regular occupation and for a number of years followed the life of a tramp. This emotional deterioration progressed more or less rapidly, depending in a measure upon the severity of the choreic movements and the extent to which the patients were prevented from employing themselves and enjoying social intercourse. In some of my cases the emotional deterioration was very moderate until the latter stages of the disease, when within a few months, it increased with such rapidity that at the end the patients presented extreme emotional apathy. Early in the development of the mental symptoms, the patients lost their sense of responsibility in reference to their own welfare and that of their family. It is a striking fact in Huntington's chorea that the patients from the onset fail to appreciate the significance of the nervous and mental symptoms, and throughout maintain an attitude of absolute indifference as to their incapacity. Many of the patients expressed a distinct hope for ultimate recovery. The only exception occurred in two cases. These patients near the onset expressed a fear of chorea which they had seen in their own parents and relatives lead to absolute dementia and ultimate death, but in both of these cases this fear disappeared within a very few months, and they became just as indifferent as the others. The emotional deterioration appears, as in dementia præcox, out of proportion to the evidences of dementia in the other fields, and thus offers a striking contrast to the dementia seen in dementia

paralytica, which by some has been considered the true picture of Huntington's chorea.

The evidence of dementia in the other fields was observed chiefly in the matter of *memory*. The impressibility of memory usually suffered first and later retentiveness. The defects of memory in most cases progressed very slowly; even in cases of twenty-five to thirty years duration, the patients still possessed a very fair memory for both recent and remote events. Indeed, it was often surprising to observe how well the patients remembered even when speech had become almost unintelligible and when emotional deterioration was profound. Sometimes during the last few months of the disease the defects of memory increased rapidly. In only three of my cases did the memory defect ever become so pronounced that the patients could not remember such well known events as the names of their parents or children, dates of births, etc.

Difficulties of *apprehension* occurred only during the late stages of the disease, the patients continuing capable of securing a pretty good grasp upon what took place about them until other evidences of dementia were far advanced. The patients also usually remained fairly well oriented until the last stages of the disease. In the field of *thought*, there regularly developed rather early evidences of paralysis of thought, that is, an increasing limitation of the association of ideas. The patients read and conversed but little, and even during conversation had comparatively little to say. While this defect developed early it did not become profound until the last stages of the disease. It is a notable fact that even in advanced paralysis of thought the patients never showed incoherence of thought similar to the desultoriness encountered in dementia præcox. Whatever knowledge they retained, they were able to render coherently.

Relative to *judgment*, the patients rather early showed certain peculiar defects, such as the lack of insight into their disease and their incapacity. The lack of insight became more striking as the disease advanced so that frequently in the end stages, even though they were bedridden and unable to feed themselves, the patients still maintained that they were able to earn their living by such manual labor as sewing, and house and farm work.

Not all cases with progressive dementia reached absolute and profound dementia before death from the disease as usually happens in dementia paralytica. Indeed, some of my cases at death presented only a moderate degree of dementia. Though dementia had progressed slowly in some cases, it advanced very rapidly in the last few months, coincident with the rapid extension and increase of the choreic movements.

The second group of mental symptoms in Huntington's chorea, which I have chosen to designate cases with crazy symptoms, occurred in fourteen cases. These cases presented two fairly well defined pictures.

The first picture is very similar to that encountered in the hebephrenic form of dementia præcox. The onset was gradual. The patients first complained of insomnia and malaise. Within a few weeks there gradually appeared delusions of reference and personal injury, moderate despondency and sometimes also some anxiety. There was no particular disturbance of thought; there were no active hallucinations, clouding of consciousness or disorientation. In female patients, particularly, delusions of infidelity were very prominent during this period. These crazy symptoms usually disappeared within a few months leaving the patients with some signs of apathy and indolence. Thence there developed the characteristic picture of simple progressive dementia already described

under the previous group. In most cases the early crazy symptoms were accompanied by some increased irritability with impulsiveness and sometimes silliness. In one case the crazy symptoms developed during childbirth and wholly disappeared by the sixth month and it was not until some months later that the characteristic symptoms of progressive dementia appeared. In this case also, religious delusions of self-condemnation and delusions of self-accusation continued for five years and accompanied the symptoms of dementia which, however, did not progress extensively until the year previous to death. In some of the cases, there later occurred exacerbations, very similar in character to those observed in hebephrenia, during which delusions, irritability, despondency and impulsiveness persisted from a few weeks to several months. Suicidal attempts during such periods of despondency were often observed. A few cases, both during the early despondency and the periodic exacerbations, expressed indefinite somatic delusions. In one case, the crazy symptoms did not develop until sixty and consisted mostly of persecutory delusions and hypochondriasis which continued until sixty-three, when they gradually disappeared with the advent of dementia. In one case the exacerbation developed two months preceding death and continued accompanied by refusal of food.

The other picture, shown by three of my cases, is most like that of the anxiety psychosis with distinct intermissions. Here, also, the onset was gradual with increasing apprehensiveness, delusions of self-accusation, of condemnation, of punishment, occasional hallucinations of hearing, considerable agitation and frequent suicidal attempts. There was no disturbance of thought or attention, or clouding of consciousness except in one case. In one case there were three such periods extending through five years

with intervals from six months to two years. Ultimately, in all of these cases the characteristic progressive dementia of Huntington's chorea, as already described, developed rapidly preceding death. In one case there occurred clouding of consciousness with stupor. In this case there were two such attacks, lasting eight and six months respectively. The evidences of progressive dementia appeared only some months after the recovery from the second attack. In only one of the twenty-eight cases did there at any time develop expansive delusions. These were of a moderate degree and lasted but a few months. As regards any relationship between the choreic and the mental symptoms, it can be said that in the greater number of cases the choreic manifestations are far more severe and produce permanent incapacity sooner than the mental defects. In a few cases, particularly during the terminal period, both the choreic symptoms and the mental symptoms advanced rapidly. In none of my cases was there observed any such abatement of choreic symptoms as is indicated by Wollenberg, during the period of dementia. On the other hand, there was one case with crazy symptoms in which both the mental and physical symptoms gradually abated and wholly disappeared for some months.

Relative to the date of onset of the mental symptoms, there were among the fourteen cases with crazy symptoms, four in which the mental symptoms antedated the development of chorea, a fact which is not generally known, though it has been pointed out by Sinkler, and a case has been reported by Hallock. In one case, that which developed during childbirth, the symptoms antedated the chorea by one year; in a second case by three years; in a third by ten years, and in the fourth case by thirteen years, the last two cases having been diagnosed as cases

of dementia præcox at the time of their first commitment to the hospital. In the ten remaining cases, the crazy symptoms developed coincidently with the choreic movements in four, while in the others, they developed from one to nineteen years later, the average being seven years after the first appearance of the choreic movements.

In the eleven cases that presented progressive dementia, the onset varied from two to ten years and averaged six years after the development of the choreic manifestations. In only one case did the progressive deterioration develop coincidently with the choreic movements. In two other cases in which there was not a sufficiently full history of the mental symptoms to classify them, it was known that the mental symptoms occurred eighteen years after the appearance of the chorea in one, and coincidently with the choreic symptoms in the other.

There is one case in which the choreic movements have existed for twenty years without the development of mental symptoms. This patient is now seventy years of age, having been choreic for twenty years. Absence of mental symptoms have been noted by Ewald, Kornilow, Huber and Söldner, but in all of these cases it is notable that the disease was of short duration. The duration of the disease must always be taken into consideration, as well as the cause of death, before the assertion can be made that a case of Huntington's chorea never had mental symptoms. In one of my cases the patient died of pneumonia at forty-five, after having had choreic movements for ten years without any other mental symptoms than slight tendency to seclusiveness and some "queerness." This observation is particularly applicable in the two cases of Huber who, he claims, were intelligent officials up to the time of their death.

REV. CHARLES OSCAR WATERS, M.D.

I. Biographic Sketch. II. Location of His Cases.

I.—It is an anomalous sounding record that we find on gathering the history of Dr. Waters. He led a life of such multiple and varied activities, though rich in experience, so discrete in detail that no person appears to have realized the thread of personality that connected it all.

From boyhood he seems to have been something of a wanderer a tendency stimulated at times by an enforced search for health. And his life purposes suggest a touch of the same; early a contributor to medical progress, and finally a devotee of ultra-orthodoxy in religion. In the earlier days of the country a few men of parts united medicine and theology, but rather from force of circumstances than duality of purpose.

His opportunities for observing these cases of chorea were brief compared to those of either of the others of this first quartet. And there is nothing to show that he ever was aware of the interest of his observations, or their rediscovery, to medicine. He was not deeply interested in the matter originally, and merely wrote what he did as a courtesy to Dr. Dunglison. The chance for really establishing a valuable and lasting contribution was allowed to pass. And medically we recall his brief description as a curious episode and for its inferential value in carrying back the known duration of one group of cases.

As to recognition, if it is to be tolled to any one here, it should go chiefly to Dunglison, who was responsible for both Waters' and Gorman's papers.

Dr. Waters was born at Franklin, Delaware County, N. Y. (western Catskills), in 1816. His father was Ozias Waters, who as a youth in 1798 migrated with his parents (Mr. and Mrs. Joseph Waters) from Hebron, Conn. The father married Miss Anna Phelps (died 1876, *act.* 82), and was in 1835 a member of the Building Committee and of the first Board of Trustees of the Delaware Literary Institute. The doctor had three sisters and one brother, Erastus. All the family have been highly regarded.

His boyhood was passed at Franklin and the adjoining town of Delhi. He tells of a visit when eight or nine years old to Cooperstown, of being present at the organization of a church at Delhi in April, 1831, of riding on the new Albany & Schenectady R. R. "a few years earlier" than 1834, and of teaching a district school near Delhi in the winter of 1833-4.

He prepared for college at the Delhi Academy, and in the fall of 1834 entered Williams. "His hair was frosted" before that. "Was present from 1834 to 1837 as Freshman Sophomore and Junior. He left before the annual catalogue of 1838 was printed. He received no degree"* (Williams College records). The Rev. Henry M. Field was a classmate, and they ever preserved "a warm and uninterrupted friendship." Referring to the class of 1838 at Williams, Waters writes: "The subscriber left * * * mainly because a kind faculty and several severe hemorrhages from the lungs admonished him he need not stand upon the order of his going.* * * He

*Honorary recognition was given him subsequently, according to Rev. Dr. Patton, President of Princeton Theological Seminary, and a special friend of Dr. Waters' later years. He says: "During my stay in Chicago, that is between 1871 and 1881, he received the degree of M.A. from Williams, for I remember very well the pleasure it gave him to receive the diploma."

left, expecting to return in a few months. He never went back. * * * His recovery was so very slow that it changed all his life plans."

In fact he left college late in 1836, and the same year took the position of assistant teacher at the new Delaware Literary Institute, "at the dazzling compensation of ten dollars and fifty cents per term for two hours daily labor." He only served the winter there and then found a better position. "It was in the spring of 1837 * * * that, from my home near its source, I floated down the Delaware on a lumber raft to Belvidere, New Jersey." There he became principal assistant to the Rev. Dr. I. N. Candee, at his private academy. Early that summer they drove over to Easton, to attend the Lafayette College commencement. Waters was taken with a chill, returned the same evening, and was nursed by the Candees through a fever that lasted most of the summer. It is an evidence of his spirited character, that although always subject to periods of invalidism he succeeded in reaching more than the allotted age, in filling out a life of great activity, and in preserving to the end a vigorous pen for his favorite topics.

He relates calling in the fall of 1837 on an enfeebled ex-president of Williams, near Newark, and evidently being influenced by him to the ministry.

"Studied for the ministry and then medicine, both of which professions he followed more or less" says Dr. White. In the winter of 1839-40 he attended his first course of lectures at the Medical Department of Columbian University, in Washington, D. C. "At that time, and under a rule of the institution, each Senator of the United States could select a student for one gratuitous course of medical lectures, and I was there as the protégé of the Hon. Silas Wright, then one of the Senators from

New York." Waters then changed to Philadelphia, graduating at Jefferson Medical College in 1841.

He never practiced medicine here at the East. "His health failed at this time" is the reason given for his going West. He reached St. Louis in the fall of 1844, and settled at Muscatine, Iowa, the same year. He speaks of this as "our first and only home in Iowa." "During the 'Mexican War' the subscriber, carefully guarding his commission as an Assistant Surgeon in the United States Army, was a passenger on the brand new steamer 'St. Paul,' on her first trip from St. Louis to New Orleans." Except this statement of his, nothing more is known of such service. About 1848 he married Miss Fannie Cummings at Dayton, Ohio. For one year he was editor of the Muscatine paper. In 1858 he was a member of the Iowa State Medical Society. He formally entered the Presbyterian ministry as a licentiate in 1860, becoming Superintendent of State Colportage in 1862.

He remained in Iowa just twenty years. He was only fifteen years in active medical practice, however, devoting himself thereafter to clerical and religious lines. His friend, the Rev. Dr. Frothingham, kindly supplies the following: "I have searched the records of the Chicago Presbytery and find only this entry concerning him that on April 11, 1866, he was 'received as a licentiate from the Presbytery of Cedar (Iowa).' He never was ordained, but as a licentiate served the Board of Publication for several years as manager of their depository here."

He became an elder in the Jefferson Presbyterian Church there, and in 1880 a Trustee of the Chicago Theological Seminary. In the great Chicago fire of 1871 his home was burned. The later years of his life he was the financial agent of the Dubuque German Theological

Seminary, and in that capacity traveled and lectured extensively through the Middle and adjacent states.

He retained a great affection for the home of his youth, revisiting it in 1864, 1875, 1881, 1883, etc. He died at Chicago, of chronic Bright's, in 1892. The only child was "one son, who died a few years ago." And his widow died about ten years since.

"He wrote frequently, but chiefly for religious papers." His niece, Miss King, says: "I was more familiar with his life as a minister than a physician." From 1859 until his death, a third of a century, he was a regular contributor to the *Presbyterian*, his last article appearing in August, 1892. He usually wrote, "Notes from the Northwest," and later, "Notes from the Interior," always signing them "Calvin." Some of the above excerpts are from that source.

II.—There are now at hand sufficient facts bearing on the location of his cases to permit a reasonable and positive conclusion.

To Dr. White, of Franklin, I am indebted for looking up Waters' history "among our oldest inhabitants." "He did not practice either profession in Franklin, so presumably wrote the article on Huntington's chorea while on a visit here, as he came here for many years frequently to visit his father." In fact it was just after finishing his course at the medical school that he sent back his description to Dunglison. "In regard to Huntington's chorea here, I have never seen a case in the eighteen years of my practice in this locality."

More directly to the point is Waters' own reference to it as "a singular affection somewhat common in the southeastern portion of the State." It was once suggested that by this expression Waters referred to Easthampton, L. I., the region of Huntington's cases, but this has a

different meaning to New Yorkers, from what might naturally be assigned to it by people elsewhere. It is customary to refer to Long Island as such. However, to make doubly sure, a native of Waters' home region was asked what such a statement would mean? He immediately replied: "By southeastern New York, I should understand Westchester County." And that is the most strict and literal interpretation to be placed on Waters' own words. This of course promptly identifies his cases as belonging to the same series described twenty-two years later by Lyon (*v. infra*).

This is borne out in two ways:

Bedford is only about a hundred miles from Franklin. Although but indirectly accessible from the latter, it was at that period a county seat and something of center; while the lack of railroads prevented even itinerants from often journeying to such an inaccessible spot as Easthampton would have been to him. As we have seen, Waters, even in his younger years, was always travelling, for study, health, teaching, or just *Wanderlust*. And one of his classmates at Jefferson, Dr. G. C. Finch, always lived in North Salem, a town next to Bedford and usually the home of some of these choreics. It is thus easy to see how he ran across cases of this group.

His actual relation to the chorea region remains enigmatic. It was likely in the blank period of his history from the fall of 1837 to that of 1839. Other times and experiences are mentioned or can be traced. But, for whatever reason, he appears to carefully avoid referring to this one matter and particular period. The present account may serve to elicit authoritative information hereon from some source.

An additional strong inference can be drawn from his mention of the word 'magrums';—"known among the

common people as 'the magrums.' Whence the name originated I do not know, but if it be a corruption of the word 'megrim,' I am at a loss to understand how it came to be applied by the vulgar to the disease of which I am speaking, and which has nothing in it analogous to ordinary hemicrania or megrim."

It may add clearness to consider the source and meaning of the word 'magrums.' Well may Waters have been puzzled if unaware of its origin. In fact none of the special or general dictionaries give anything regarding it (except Dunglison's Medical, edts. 1844-60, which was merely following Waters), and its ultimate derivation may be left to the philologist. It is, however, a household word long in use up and down the Hudson River valley, especially amongst the Dutch and those in touch with Dutch influence. Although it has thus scattered much further, it is stated by those most familiar with it to be a Dutch word (not in Kalisch's Dutch-Eng. Dict.). Neither in its origin nor in its significance in common use has it anything to do with megrim, megrims or migraine. It is used to imply restlessness or nervousness, about equal to our "fidgets." Children who do not keep quiet are told they have "the magrums." If a person is kept awake by cramps in the legs, it is said to be owing to the "magrums." It was a natural designation to apply to this affection. But it would only be introduced where the Dutch influence was strong. Of course to the non-Dutch in the chorea region the word would come to have a more or less specific meaning. Formerly at least, this term does not appear to have been thus used about East Hampton, where the chorea was referred to as "that disorder" or as St. Anthony's dance (personal communication of Dr. Huntington). Consequently, as between the English-settled, eastern portion of Long Island, and the partly Dutch-

settled region above Manhattan (these being the seats of the only groups of original cases that come in question), the latter is alone probable.

There are thus three reasons for and none against this location of Waters' observations. It serves to carry back the heredity in Lyon's cases twenty odd years. And it may fairly be concluded that this group has now existed for three-quarters of a century and doubtless far longer.

The Editor.

DR. CHARLES ROLLIN GORMAN.*

I. Personal Sketch. II. His Relation to the Chorea Question.

I.—Dr. Gorman was born August 4, 1817, at Barkhamsted, Conn. His boyhood was passed there, and he is even said to have taught school in Connecticut. His last teaching was at Pittston, just before or while studying medicine. His father was Dr. James T. Gorman, who sometime in middle life moved with his family from Connecticut to Benton, Pa. (then in Luzerne County, but becoming about 1878 a part of the new Lackawanna County), where he practiced until his death in 1861. It is thus evident that the younger Gorman, like Huntington, had the advantage of medical ancestry, in preparing his paper. The paternal grandfather was "an Irish school-master named O'Gorman." Lee's *Barkhamsted*, 1881, mentions both Drs. Gorman as natives of that town.

Dr. C. R. Gorman's mother was Lois Beecher (Gorman), a second cousin of Henry Ward Beecher, and a daughter of Dr. Amos Beecher, for half a century the physician of Barkhamsted.

*The Editor is especially indebted to Dr. L. H. Taylor, of Wilkes-Barre, for information regarding Dr. Gorman.

The younger Dr. Gorman graduated from Jefferson Medical College in 1848. His preceptor was Dr. Throop, of Scranton. He practiced in Phillipsburg, N. J., for twelve or eighteen months. "Later went to Burlington, N. J., but I think did not follow his profession there as he delivered a course of lectures in chemistry in the towns around Philadelphia. * * * Later settled in Pittston, Penn, where he continued his practice to the time of his death. He was at one time Postmaster of Pittston and a member of the House of Representatives of Pennsylvania. He was also a member of the Odd Fellows and was Past Master in St. John's Masonic Lodge, of Pittston. He was the first President of the Pittston Library Association and Vestryman in St. James' Episcopal Church." He became a member of the Luzerne County Medical Society in January, 1863; and was also interested in pharmacy.

He was twice married. The first wife was Mehitable Schooley, who had two children. The second wife was Annette Jenkins; the widow and their one child, Miss A. J. Gorman, still reside at Pittston.

Dr. Gorman died April 3, 1879.

II.—Gorman's relation to the chorea question.

The following item is given in Dunglison's *Practice*, 3 ed., 1848, p. 218: "In an inaugural dissertation, presented before the Faculty of Jefferson Medical College, of Philadelphia, by Dr. Charles R. Gorman, of Luzerne County, Pa., the writer states that this affection prevails also in other portions of the country. According to him, it seems to be circumscribed by neighborhood boundaries, and to be confined to sections of the country, the inhabitants of which are intimately connected in their social or business relations. 'May not this circumstance,' he asks, 'sanction the inference that the cause exists in the

influence the *moral* is known to exert over the *physical*—the sympathy of imitation?" "

This paper it has not been possible to trace further. Dr. Dercum kindly reports: "The Thesis of Gorman in 1848 has been lost with some other material at the tearing down of the old College Building." And from the family: "In regard to the thesis we have not been able to trace it."

The above excerpt then includes all that is preserved of Gorman's paper. From that alone it would be impossible to say whether he was really describing this form of trouble or not—although with Dunglison's endorsement it may be accepted that he was.

It has been possible, however, to establish the location of Gorman's cases, and thereby as well to verify the correctness of his work.

That his cases were not from his early Connecticut associations is variously shown. His description implies Pennsylvania; his birthplace was a long distance from the chorea-section of Connecticut; and Dr. Howd, of Winsted, writes, "I have been acquainted in Barkhamsted and Barkhamsted Hollow for years, but never knew of a case of Huntington's chorea."

On the other hand, the Pennsylvania side is positive. In the well-known article by Sinkler (*Jour. Nerv. and Ment. Dis.*, 1889, p. 86), the S— family, from which his Table II was compiled, belonged at Nicholson, Wyoming County, Pa. And this town directly adjoins Benton in Lackawanna County, the place, as we have seen, where Gorman's father practiced medicine, and where the junior himself naturally passed much time in his younger years. In fact, the active centers of the two towns are only a couple of miles apart. Moreover, in 1888 a great-grand-daughter of the first case was sixty-seven years old,

showing that the group must have reached back far beyond Gorman's time. It is therefore certain that Gorman in his younger years had ample opportunity to see real cases of this trouble.

This may very suitably be termed the Wyoming Group. It may provisionally be considered the third of the older groups of cases (I, East Hampton; II, Bedford; III, Wyoming Groups). It has consequently been utilized at least twice and quite independently in descriptions.

The Editor.

IRVING WHITALL LYON, M.D.

I. Personal Sketch. II. Location of his Cases.

I.—Dr. Lyon was born October 18, 1840, at Bedford, Westchester County, N. Y. His father was Solomon R. Lyon, a ruling Elder in the local Presbyterian Church in his later years, dying in 1868. The doctor "attended the Public School in Bedford in his boyhood," and gained his further preliminary education at an academy in the town, between 1855 and 1860. Received his M.D. from the University of Vermont, in 1862, and from the College of Physicians and Surgeons (N. Y.), in 1863. At the Vermont school, "he was credited with coming from Bedford, N. Y., and his preceptor was O. W. Peck."

He was Acting Assistant Surgeon, U. S. A., in June and July, 1862. Became Demonstrator of Anatomy, 1862, in the then famous Berkshire Medical School. Served as a member of the house staff, Bellevue Hospital, New York City, 1863 to 1864. It was while occupying this position that he published the article on chorea.

In the fall of 1864 he settled in Hartford, Conn., and there remained. He was Medical Director of the Hart-

ford Life and Annuity Insurance Company from the time of its incorporation in 1868 until his death. He was a member of the Connecticut Medical Society, the Connecticut Historical Society, Society of Sons of the American Revolution, and was President of the Hartford County Medical Association at the time of his death.

"Dr. Lyon contributed articles to the New York and Philadelphia journals and to the Hartford Medical Society, and published several monographs. For the last twenty years of his life he devoted his leisure to anti-quarian studies. In 1891 he published a volume entitled 'The Colonial Furniture of New England.'" (Not to be confounded with I. W. Lyon, a D.D.S. of New York City.)

He died March 4, 1896, of pneumonia, and was buried at Bedford, "in the Cemetery in which lie his ancestors." He left a wife and three children. Dr. Irving P. Lyon, Librarian at the University of Buffalo, is his son. Lyon's obituary in the *Proceedings of the Connecticut Medical Society*, 1896, page 327, gives no intimation of his relation to the chorea question.

II.—It is now possible to determine exactly the location of Lyon's cases. His article begins as follows: "The writer has been familiar from childhood with a type of chorea so unlike in its *origin* to anything described in our standard text-books." While he avoids specifying the region of his cases more exactly, even dating his article Bellevue Hospital, he does note that two were in the State of New York, and one in Connecticut. As we now know that he was born at Bedford, Westchester County, New York, adjoining the western boundary of Connecticut, spent his boyhood there, and made that his home until taking his medical degree, it is evident that his observations were made in that region.

That there were cases of this kind in southwestern Connecticut (the part of Greenwich directly adjoining Westchester County, N. Y.; in fact Bedford was in early days a part of Connecticut), and even over the border in New York State, has been known to the writer for many years, though not aware until this search that this was the site of Lyon's observations. Such cases it was said had existed thereabouts for more than fifty years. There it is always known as "magrums."

Further corroboration is furnished by Dr. L. H. Miller, a native of Bedford, who not only prepared for college there but later taught at the academy. He states that several families with this affection have lived in the poorer eastern part of that town, that cases still exist thereabouts, and that "up there they always call it 'magrums.'"

From this surfeit of facts* it can be positively stated that Lyon's cases were from the Bedford region in New York and the adjacent part of Connecticut. This also establishes a definite center about which one of the groups of these cases has existed for a long period. For convenience it can very properly be termed the Bedford Group (*v. also supra*, sketch of Dr. Waters).

The ear-mark of cases in this group appears to be their popular local designation as "magrums," a term which Lyon naturally medicalized into "megrimms."

The Editor.

*A recent letter from Dr. O. W. Peck, now of Oneonta, N. Y., says: "I had the honor of being the preceptor of the late Dr. Irving W. Lyon. * * * He took up the study of chorea, however, on his own account without assistance from me. * * * When I began practice in Bedford, Westchester County, forty odd years ago—before the war—the disease—magrums the people generally called it—was quite prevalent in that vicinity extending over the state line into Fairfield County, Conn. To some extent, as I remember, it was regarded as hereditary."

NOTE ON THE TEMPORAL, GEOGRAPHIC, AND RACIAL DISTRIBUTION OF HUNTINGTON'S CHOREA.

The wide occurrence of this form of chorea has been noted by several writers (Schlesinger, King, *et al.*). And the appended bibliography warrants a further word, showing as it does its almost limitless range.

How far back in time its existence can be traced has received little attention. Meynert, of Vienna (1885), speaks of a family in which it had occurred for a hundred years. Huntington carries back the Easthampton group nearly as long, and on the more positive medical basis of his grandfather's observation. Tilney shows (*v. supra*) in agreement with Jelliffe, that in this country one line has existed from well back in the colonial period, and yet with no indication of being nearer its source or of any variation in the "strain." Neither the ordinary taints, as noted by Huntington, nor yet the biblical third and fourth generations measure the length of this shadow when once evoked. Much less than paresis is it to be considered an affection of modern times merely. From the lack of any known etiology but heredity, it might be expected to reach back indefinitely.

On the other hand, so far as anything has been discovered, there is a surprising absence of historical reference to this condition. And cases, such as one given by Strümpell above, suggest that it may occasionally originate *de novo* (not becoming a family affair unless in one young enough to transmit it).

An old Easthampton tradition, as personally related by Dr. Huntington, is to the effect that this disorder came as a curse upon those who persecuted Roger Will-

iams. While this can hardly be true,* it may have originated in some terrible shock, privation, or combination of experiences such as the early immigrants and settlers must have had in plenty.

While consequently the origin of the older American "strain" has not been reached, except only that the mother line appears to have cropped out first at Stratford, Conn., the farther back it can be traced the more probable is it that the choreic vein was brought over by one or more early settlers and has merely persisted.

One way may be indicated by which this "strain" can be traced still further back, *i. e.*, if it existed earlier. It is only possible, however, provided choreic cases can be found in one or more of the collateral lines of descent. If such are found and are not of later introduction by marriage, then it will be evident that the strain existed at least as far back as the point where the lines branched from the parent stem. Whether such diathetic heredity constitutes or depends on a Mendelian unit does not as yet appear to have been definitely settled.

Letters, records, and printed scraps relating to these families might be expected to contain something hereon. Historical reference hereto may yet exist. And even family traditions in the respective localities have not yet been carefully gathered. It is easy but not warrantable to assume that progress in carrying back our knowledge of this matter is impossible. As the search has a biological and ethnic as well as purely medical importance, it may be hoped that the retrograde phase, will be better exploited.

On the geographical side it is to be noted that cases have been described from every country in Europe (unless possibly Turkey and Norway-Sweden be excepted),

*It has a certain value however, implying that the trouble was not of local origin but was brought by early settlers from the mainland.

including of course England, from the West Indies and various points in South America, as well as from Canada and a large number of states in all parts of the Union. This means that it is co-existent with European civilization at least. On the whole, it may possibly be more frequent in southern than in extreme northern Europe; at any rate the Italians have been among the larger contributors to the study of the subject.

That it is not, however, limited to any one order of culture is shown in various ways: In America at least, the locations of the older group-centers do not suggest that it is in any way a product of the so-called strain of modern life; if so at all it must have been on the part of some of their pre-choreic forbears.

It is certain also that there is no color or race boundary. Mills and Burr (1890) included one case in a negro family; Dercum (1891) also gave such a case; and Drewry (1895) added another. If peoples as different as the Caucasian and the Ethiopian can suffer therefrom, it must be racially of universal occurrence.

It is for one reason this wide distribution geographic and racial and the consequent frequency of cases that has had much to do with the leading position this disease has assumed in the study of hereditary neuroses.

The Editor.

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In all, over two hundred articles and original contributions hereon have appeared, besides a large additional amount of allied literature in text-books, treatises, discussions, and the differential diagnosis from athetosis, tic, myoclonia, etc.

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